

# ANOMALOUS PULMONARY VENOUS DRAINAGE OF THE RIGHT LUNG INTO THE INFERIOR VENA CAVA

BY

ROBERT L. FRYE, HIRAM W. MARSHALL, OWINGS W. KINCAID,  
AND HOWARD B. BURCHELL

*From Section of Medicine, Mayo Clinic and Mayo Foundation Rochester, Minnesota, U.S.A.*

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Of the various non-obstructive anomalies of partial pulmonary venous drainage, that of a large pulmonary vein draining the right lung into the inferior vena cava above the liver is one most deserving the designation of a syndrome. The first reports of this anomaly included descriptions of associated abnormalities such as dextroposition of the heart and a systemic arterial vessel supplying the right lower lobe (Cooper, 1836; Park, 1912). From the appearance of the anomalous pulmonary venous trunk draining the right lung, the term "scimitar syndrome" has been suggested (Neill *et al.*, 1960). Five cases studied at the Mayo Clinic have been chosen to demonstrate the following variations in the clinical picture: *normal right lung or partial agenesis thereof; normal heart or dextroposition thereof; absence or possible presence of an anomalous systemic artery to the right lung; and absence or presence of an associated atrial septal defect.* In addition, characteristic features of the splitting of the second heart sound in patients with partial anomalous pulmonary venous drainage and intact atrial septum are presented.

Ten patients with anomalous pulmonary venous drainage of the right lung into the suprahepatic portion of the inferior vena cava have been examined at the clinic, five of whom have been selected for review. Cardiac catheterization was performed in three of the patients, using techniques described previously (Symposium on cardiac catheterization, 1953), and the phonocardiograms were recorded by means of a Sanborn twin-channel phonocardiogram.

## CASE REPORTS

*Case 1. Anomalous pulmonary venous drainage of the right lung [APVDRL] into the suprahepatic inferior vena cava without associated abnormalities.* A girl, aged 11 years, was seen at the clinic for evaluation of mental deficiency and hypothyroidism: there was no history of any cardiovascular or respiratory symptoms. Examination revealed an intelligence quotient of 64, but thyroid function was normal. A grade 2 systolic murmur was present in the second left interspace. An electrocardiogram was within normal limits. Radiography of the chest showed an anomalous vascular shadow in the right lower lung field and tomograms demonstrated that the vessel descended toward the diaphragm along the right cardiac border. No further studies were made, and the patient has remained entirely asymptomatic during the seven years since the first examination. Follow-up radiographs have shown no change in the size of the heart.

*Case 2. APVDRL into the suprahepatic inferior vena cava with dextroposition of the heart and agenesis of the right upper lobe.* A woman, aged 45 years, was first seen at the clinic because of nervousness: she had no cardiovascular or respiratory symptoms. On examination, however, it was noted that the heart sounds were

loudest to the right of the sternum, with a prominent right parasternal lift. Radiographs of the chest revealed dextroposition of the heart with loss of volume of the right lung, in addition to a large vascular shadow along the right cardiac border. The electrocardiogram (Fig. 1) is consistent with dextroposition of the heart in contrast with a mirror-image dextrocardia. Because of the abnormal radiographic appearance of the right lung, bronchoscopy and bronchography were performed. These studies revealed that the bronchus of the right upper lobe terminated about 1.5 cm. from its origin and that the lung at the apex of the thorax was supplied by the bronchus of the right middle lobe. This pattern suggests agenesis of the right upper lobe. One of the films obtained at the time of bronchography is shown in Fig. 2. Cardiac catheterization was performed and revealed an increase in oxygen saturation in the suprahepatic portion of the inferior vena cava consistent with a left-to-right shunt. The pulmonary arterial pressure was normal, and the magnitude of the shunt was 28 per cent, with a pulmonary index of  $4.6 \text{ l./min./m.}^2$

*Case 3. APVDRL into the suprahepatic inferior vena cava with normal splitting of the second heart sound and possible presence of an anomalous systemic arterial supply to the right lower lobe.* A girl, aged 15 years, had been noted to have a typical "scimitar shadow" on routine radiography of the chest. There was no history of any cardiac or respiratory symptoms. On physical examination she was found to have a grade 2 systolic murmur in the second and third intercostal spaces on the left with normal splitting of the second heart sound. This is illustrated in Fig. 3 with a phonocardiogram recorded in the second left interspace. It may be noted that with inspiration the interval between the aortic and pulmonary components of the second heart sound is 0.04 second, and during expiration the two components are fused. It can also be seen that the interval between the R wave of the electrocardiogram and the aortic component of the second heart sound (R-A2 interval) is 0.33 second in inspiration increasing to 0.35 second during expiration. In the lower half of Fig. 3 the phonocardiogram is recorded simultaneously with the carotid pulse. This demonstrates that the duration of mechanical systole, measured from the upstroke of the carotid pulse to the dicrotic notch, decreases during inspiration and increases during expiration. This movement of the aortic component of the second heart sound is normal and suggests that the atrial septum is intact. At the time of cardiac catheterization, a left-to-right shunt into the suprahepatic portion of the inferior vena cava was demonstrated. The magnitude of the shunt was 42 per cent, with a systolic

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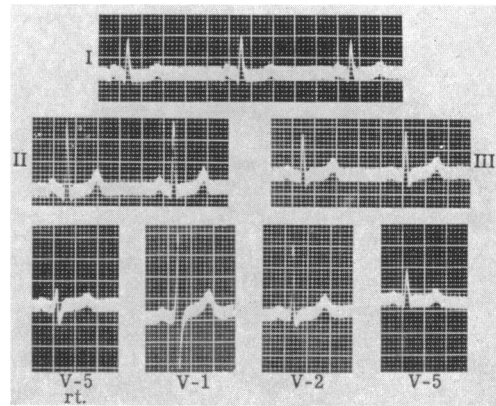


FIG. 1.—Case 2. Electrocardiogram illustrating upright complexes in lead I and a right ventricular type of complex in V-5 rt.

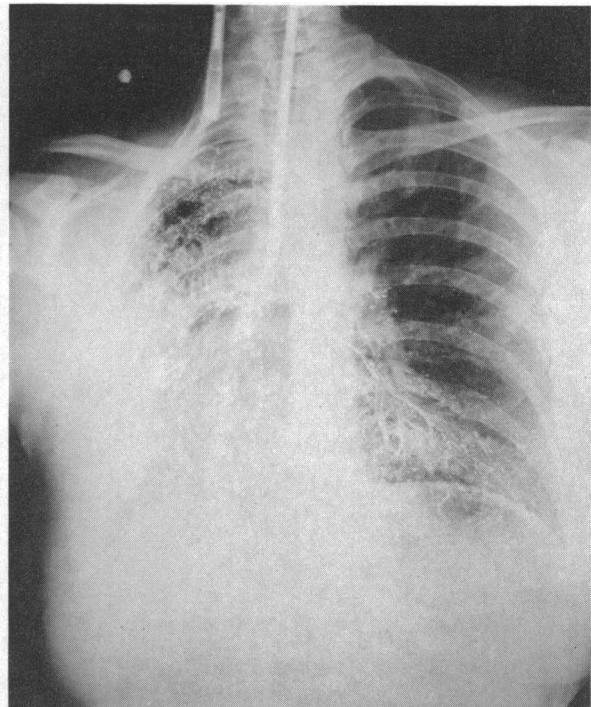


FIG. 2.—Case 2. Bronchograms illustrating termination of the bronchus of the right upper lobe about 1.5 cm. from its origin. The apex of the right lung appears to be supplied by the bronchus of the right middle lobe, suggesting agenesis of the right upper lobe.

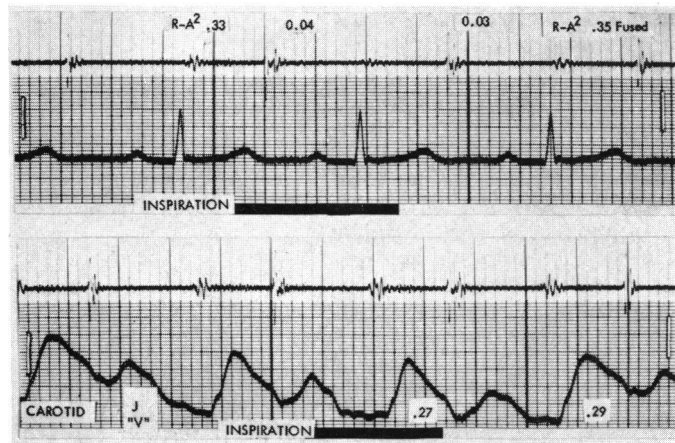


FIG. 3.—Case 3. Phonocardiogram recorded in second left interspace, with electrocardiogram in upper half of figure and carotid pulse in lower half. Note fusion of the two components of second heart sound in expiration and normal increase in duration of mechanical systole during expiration (0.27 to 0.29 sec.).

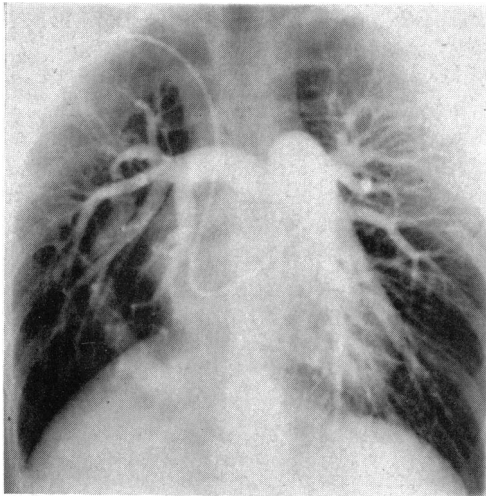


FIG. 4.—Early phase of pulmonary arteriogram of Case 3 showing diminished vascularity of the lower zone of the right lung and an anomalous vessel coursing down the right middle and lower zones.

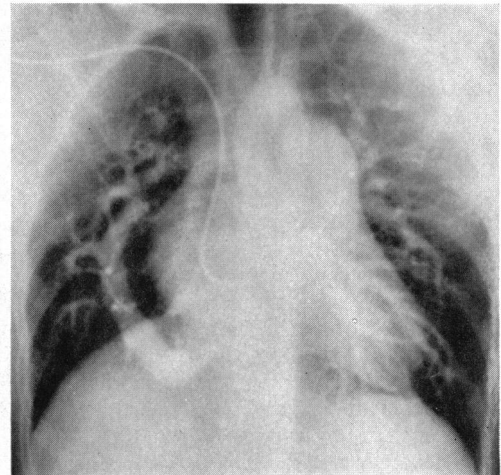


FIG. 5.—Late phase of pulmonary arteriogram of Case 3 showing the anomalous pulmonary venous trunk filled with contrast medium coursing along the right heart border towards the diaphragm.

pulmonary arterial pressure of 56 mm. and a diastolic pressure of 26 mm. Hg. An angiocardigram was performed with injection of 73 ml. of 76 per cent renovist into the main pulmonary artery. Fig. 4 shows filling of both pulmonary arteries. The vascularity of the right lower lung field is less than that on the left. This suggests the presence of an anomalous systemic artery supplying the right lower lobe; however, such a vessel could not be demonstrated with certainty. Fig. 5 shows the anomalous pulmonary venous trunk coursing along the right cardiac border toward the diaphragm.

*Case 4. APVDRL into the suprahepatic inferior vena cava with normal splitting of the second heart sound and an intact atrial septum demonstrated by angiocardigraphy.* This boy was first seen at the clinic at the age of 7 years for evaluation of a heart murmur. The child had always noted diminished tolerance to exercise, but no other significant symptoms had been experienced. On examination he appeared poorly developed. Cardiac dullness extended to the right of the sternum. A grade 2 systolic murmur was present with maximal intensity between the left sternal border and the apex. He returned at the age of 18 with continued limitation of exercise tolerance. The grade 2 systolic murmur was present as noted previously, and a phonocardiogram demonstrated normal splitting of the second heart sound (Fig. 6). A chest radiograph revealed an anomalous pulmonary venous trunk in the right lower lung field. An electrocardiogram showed a vertical axis and right ventricular hypertrophy. Cardiac catheterization demonstrated a left-to-right shunt of 50 per cent into the suprahepatic portion of the inferior vena cava, with a pulmonary index of  $7.4 \text{ l./min/m}^2$ . The pulmonary arterial pressure was normal. Selective angiocardigraphy was performed with injection of 30 ml. of 90 per cent diatrizoate sodium (hypaque) into the right pulmonary artery and also into the left pulmonary artery. Fig. 7 shows the phase of left heart filling after injection into the left pulmonary artery. The atrial septum is well outlined and appears to be intact.

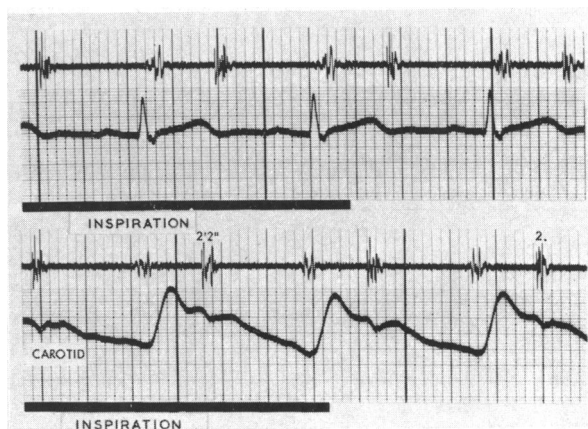


FIG. 6. Case 4. Phonocardiogram recorded in second left interspace, with electrocardiogram in upper half of figure and carotid pulse in lower half. Note normal movement of the two components of second heart sound.

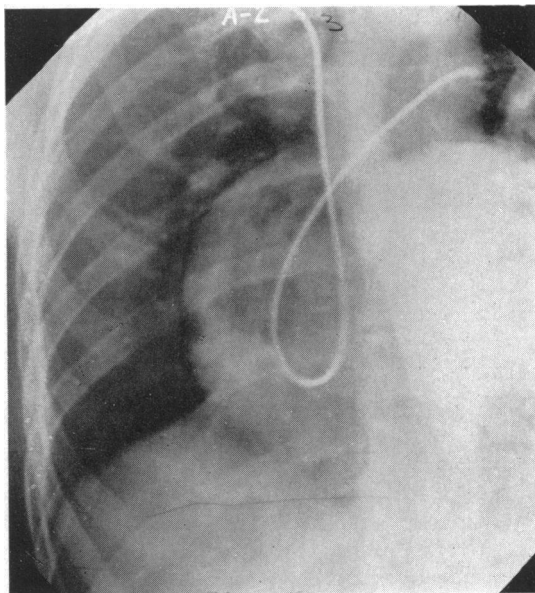


FIG. 7.—Case 4. Angiocardigram after injection of contrast medium into left pulmonary artery. The left atrium is opacified, and the septum can be seen, but there is no contrast medium in the right atrium, suggesting that the septum is intact.

*Case 5. APVDRL into the suprahepatic inferior vena cava with atrial septal defect.* A woman, 22 years of age, was first seen at the clinic because of a basal systolic murmur; she had no cardiovascular symptoms. A definite diagnosis was not established. She returned nine years later because of the recent onset of congestive heart failure and evidence of pericarditis with effusion. Cardiac catheterization revealed a left-to-right shunt of 46 per cent at the atrial level with a pulmonary arterial pressure of 50/25 mm. Hg. She returned eight months later after recovering from the pericarditis. A further cardiac catheterization showed no evidence of pulmonary hypertension, but the left-to-right shunt of 46 per cent was still present. Radiographs of the

chest showed a shadow in the right cardiophrenic angle which suggested an anomalous pulmonary vein. Operation was performed under hypothermia by Dr. J. W. Kirklin, of the section of surgery of this clinic. An atrial septal defect measuring  $2 \times 2.5$  cm. was found in addition to anomalous drainage of the right lung into the suprahepatic portion of the inferior vena cava. Complete repair was accomplished, and the patient has done well since operation.

#### DISCUSSION

The early studies of Brown (1913) provide a background for understanding the embryology of anomalous drainage of the right lung into the inferior vena cava. The origin of the lung from the primitive foregut results in anastomoses between the ultimate pulmonary venous and splanchnic circulations, the persistence of which may result in the clinical syndrome under discussion. It has been well established that this anomaly may be associated with a variety of abnormalities of the right lung and bronchial system. Halasz *et al.* (1956) described a mirror image of the left lung on the right side and cystic diverticula of the bronchial tree. Actual bronchiectasis has also been noted (Drake and Lynch, 1950). However, probably the most common abnormality is hypoplasia or agenesis of a portion of the right lung. Although patients with such pulmonary anomalies may be asymptomatic, others have been reported with repeated pulmonary infection (Neill *et al.*, 1960).

The essential haemodynamic abnormality is a left-to-right shunt. Of interest is the apparent absence of any facilitation of flow to the right lung as might be expected theoretically by the difference in the levels of pressure in the right and left atria. Studies of differential oxygen uptake by the lungs were not performed in the present group of patients; however, such analyses have been made by others, with conflicting conclusions. Arvidsson (1954) reported a patient in whom such studies revealed that the oxygen uptake of the right lung was only 138 l./min. as compared with 212 l./min. in the left lung. By contrast with these findings, Cooke *et al.* (1951) reported similar studies in one patient, which showed that 49 per cent of the total oxygen consumption was occurring in the right lung with the anomalous pulmonary venous drainage. Pulmonary hypertension may be associated with the large left-to-right shunt as demonstrated in one of the patients in the present study, or the elevation of pulmonary arterial pressure may be related to an increase in pulmonary resistance, with little or no left-to-right shunt, as reported by Neill *et al.* (1960).

An interesting finding in patients with partial anomalous pulmonary venous drainage in the presence of an intact atrial septum is normal splitting of the second heart sound. This occurs in spite of large pulmonary blood flows comparable to those in patients with atrial septal defect in whom "fixed splitting" of the second heart sound is present. Respiratory variation in the interval between the first heart sound and the aortic component of the second heart sound is probably the basis for this difference. Such movement of the aortic component of the second heart sound with respiration was first demonstrated by Boyer and Chisolm (1958*b*), and more recently by others (Castle and Jones, 1961). Shafter (1960) has emphasized the necessity of an intact atrial septum for this phenomenon, and he also noted normal splitting of the second heart sound in patients with partial anomalous pulmonary venous drainage with an intact atrial septum. Thus, in the presence of an atrial septal defect, the persistence of wide splitting of the second heart sound in expiration is related not only to delay in closure of the pulmonary valve, but also to diminution in left ventricular filling, resulting in earlier closure of the aortic valve (Shafter, 1960; Boyer and Chisolm, 1958*a*). The early closure of the aortic valve in expiration is the opposite of what is noted during expiration in the presence of an intact atrial septum. Therefore, in a patient who appears to have findings consistent with an atrial septal defect except for the presence of normal splitting of the second heart sound, anomalous pulmonary venous drainage should be considered.

More objective techniques are available to determine if the atrial septum is intact in such patients with anomalous pulmonary venous drainage of the right lung. Indicator-dilution curves may be helpful when obtained with arterial sampling after separate injections of dye into the left and right pulmonary arteries. In patients with an intact atrial septum, a curve recorded after injection into

the left pulmonary artery is normal in contrast with the curve recorded after injection into the right pulmonary artery. The latter shows delayed appearance, markedly diminished maximal concentration, and prolongation of the disappearance slope consistent with a left-to-right shunt distal to the site of injection. By contrast, patients with an associated atrial septal defect will show evidence of a left-to-right shunt distal to the site of injection in the left pulmonary artery. Representative dye curves from two of the patients in the present report, one of whom had an associated atrial septal defect, are illustrated in Fig. 8.

Several factors must be considered in regard to the question of advising surgical treatment for the isolated partial anomalous pulmonary venous drainage. Demonstration of a large pulmonary flow, particularly with moderate elevation of the pulmonary artery pressure in association with the

DEMONSTRATION BY RADIAL ARTERY DILUTION CURVES OF ANOMALOUS CONNECTION  
OF THE RIGHT LUNG TO THE RIGHT HEART IN 2 PATIENTS

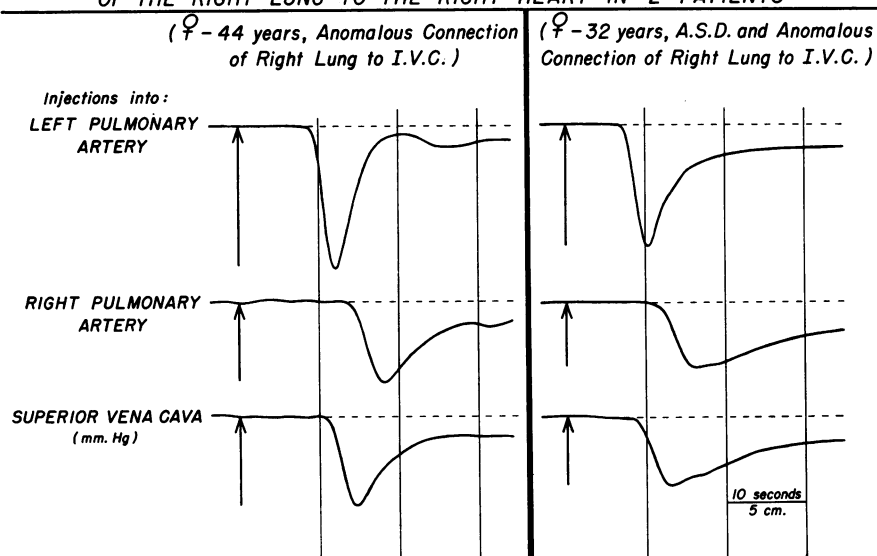


FIG. 8.—Cases 2 and 5. Indicator dilution curves demonstrating application of this method to determine if atrial septum is intact (I.V.C.=inferior vena cava; A.S.D.=atrial septal defect). See text for details.

large left-to-right shunt, constitutes a clear indication for operation. Although it is true that many such patients with large shunts may be asymptomatic, the history of this malformation is not really known. However, it seems reasonable to consider these patients as having essentially the same prognosis in regard to hæmodynamic changes as do patients with atrial septal defect. Thus, operation should be considered in order to prevent heart failure or pulmonary vascular changes as a result of the large pulmonary flow later in life. In addition, the possible consequences of extensive pneumonia, of pneumothorax, or of operation involving the left lung must be considered. Such an event would seriously compromise the patient's only direct source of oxygenated blood. Therefore, an operation seems advisable, in properly selected cases, to transpose the anomalous pulmonary vessel to the left atrium.

#### SUMMARY

Five patients with anomalous pulmonary venous drainage of the right lung into the suprahepatic portion of the inferior vena cava have been presented.

Associated anomalies may include agenesis of a part of the right lung, atrial septal defect, systemic arterial supply to the right lung, and dextroposition of the heart.

Discussion of the character of the second heart sound in patients with partial anomalous pulmonary venous drainage and an intact atrial septum is presented. Normal splitting of the second sound in a patient who has findings otherwise consistent with an atrial septal defect should suggest consideration of anomalous pulmonary venous drainage.

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